



Swansea University
Prifysgol Abertawe



Cronfa - Swansea University Open Access Repository

This is an author produced version of a paper published in :
British Journal of Oral and Maxillofacial Surgery

Cronfa URL for this paper:

<http://cronfa.swan.ac.uk/Record/cronfa19922>

Paper:

Whitaker, I., Koron, S., Oliver, D. & Jani, P. (2003). Effective management of the airway in the Pierre Robin syndrome using a modified nasopharyngeal tube and pulse oximetry. *British Journal of Oral and Maxillofacial Surgery*, 41(4), 272-274.

[http://dx.doi.org/10.1016/S0266-4356\(03\)00100-1](http://dx.doi.org/10.1016/S0266-4356(03)00100-1)

This article is brought to you by Swansea University. Any person downloading material is agreeing to abide by the terms of the repository licence. Authors are personally responsible for adhering to publisher restrictions or conditions. When uploading content they are required to comply with their publisher agreement and the SHERPA RoMEO database to judge whether or not it is copyright safe to add this version of the paper to this repository.

<http://www.swansea.ac.uk/iss/researchsupport/cronfa-support/>

TECHNICAL NOTE

Effective management of the airway in the Pierre Robin syndrome using a modified nasopharyngeal tube and pulse oximetry

I. S. Whitaker, * S. Koron, † D. W. Oliver, ‡ P. Jani §

**Demonstrator in Anatomy, Department of Anatomy, University of Cambridge, Cambridge, UK; †Senior House Officer, Department of Oral and Maxillofacial Surgery, South Manchester University Hospitals NHS Trust, Manchester, UK; ‡Specialist Registrar, Department of Plastic and Reconstructive Surgery, Radcliffe Infirmary, Oxford, UK; §Consultant ENT and Head and Neck Surgeon, Addenbrooke's Hospital, Cambridge, UK*

SUMMARY. Pierre Robin syndrome is a rare condition, first described by the French stomatologist, Pierre Robin in 1923.¹³ The severity of the syndrome varies widely, and associated upper airway obstruction is the most difficult problem to overcome. We present a case in which the airway was successfully managed with a modified naso-pharyngeal tube in addition to pulse oximetry. We also present a literature review discussing management of the airway in the Pierre Robin syndrome. The simple technique presented allows earlier patient discharge with home monitoring equipment.

© 2003 The British Association of Oral and Maxillofacial Surgeons. Published by Elsevier Science Ltd. All rights reserved.

INTRODUCTION

The Pierre Robin syndrome is characterised by micrognathia (small jaw), retrognathia (posterior displacement of the chin), glossoptosis (the tongue falls backwards) and cleft soft palate, and the incidence is reported to be 1:8500. The severity of the syndrome, which presents in the neonatal period with upper airway obstruction and feeding difficulties, varies widely. Upper airway obstruction in association with the Pierre Robin syndrome presents at, or shortly after birth as a result of retrognathia and subsequent posterior position of the tongue. The airway can be managed in a number of ways from postural nursing with the infant prone, to tracheostomy. There is much controversy regarding the long-term management of airway obstruction in children with the Pierre Robin syndrome.¹

The use of a nasopharyngeal tube to relieve high upper airway obstruction has been described in children with Pierre Robin syndrome but has not been universally successful.² Nevertheless, nasopharyngeal tubes are recommended initially to alleviate the immediate consequences of hypoxia.³ A modified nasopharyngeal tube has been recently described that does not add dead space and resistance to the airway, is well tolerated, and allows simultaneous use of oxygen prongs if necessary.⁴

We describe a patient in whom this technique was successful, and allowed early discharge with home monitoring equipment.

CASE REPORT

A female neonate, gestational age 38 weeks, was admitted to the special care baby unit with respiratory distress 1 hour after a normal vaginal delivery. She was nursed prone or on her side to stop her tongue falling back and obstructing the airway. She was monitored continuously with a pulse oximeter to alert the staff to desaturating episodes and to assess their frequency. Despite adequate postural nursing, she continued to have periods of airway obstruction, but these were short-lived. The patient was discharged after 10 days on the special care baby unit and 7 days in the children's ward, with a pulse oximeter for home monitoring of oxygen saturation. Her mother was taught how to use a pulse oximeter and told to be vigilant.

The same evening, the infant was readmitted, as her father said that the saturation had dropped by 30% on several occasions. Episodes of desaturation continued on the children's ward. So it was decided to insert a modified PVC (PORTEX) nasopharyngeal tube (size 3) into the right nostril to 6 cm. Oxygen saturation was 100% in air when she was supine immediately after the procedure. However, episodes of desaturation were continuing and it was decided to insert a larger (size 4) hard tube into the right nostril, with considerable improvement in oxygen saturation.

The benefit of the nasopharyngeal tube is shown in Table 1. Time spent with a saturation of <89% O₂ was

Table 1 Oxygen saturation after insertion of the nasopharyngeal tube

Day	Percentage of time spent <89% O ₂	Number of episodes/hour	Lowest saturation (% O ₂)	Airway management
1	8	21	60	Postural nursing
2	3	21	76	Soft size 3 tube
3	3	11	63	Hard size 4 tube
6	3	11	61	Hard size 4 tube
7	3	11	65	Hard size 4 tube

reduced by half. Three days later, she was discharged with the nasopharyngeal tube in place, and an outpatient follow up appointment two weeks later.

Modifications to the nasopharyngeal tube⁵

The required length of tube is estimated using the distance from the lateral nostril to the tragus of the ear on the same side. A tube may then be placed in the nasopharynx, temporarily taped, and its position assessed radiographically. Ideally, the nasopharyngeal tube should sit just superior to the epiglottis to relieve the obstructive breathing pattern. The length of tube at the nostril should be recorded when the correct position is established. The modified nasopharyngeal tube is prepared by cutting the measured length plus 5 cm. The tube is then cut down the middle of the underside to the measured length. Two cuts are made to each side of the midline to create two thin strips, each 3 mm. These thin strips can be used to anchor the tube to the child’s cheeks. A further cut is made to create two additional strips on the top. One of these strips is cut off at the measured length, and the other is cut off at the measured length plus 2 cm.

The nasopharyngeal tube is inserted into the selected nostril and the side strips (measured length plus 5 cm) are anchored on the cheeks on a protective hydrocolloid dressing (e.g. DuoDERM), with adhesive tape. The shorter top strip (measured length plus 2 cm) is passed over the ala and anchored on the lateral nasal aspect.

The nasopharyngeal tube is changed every 2–4 days for the first 10 days and then every 5–7 days thereafter, alternating between the nostrils if possible.

DISCUSSION

There is a much controversy about the long-term management of airway obstruction in children with Pierre Robin syndrome and delays in securing the airway at birth has been documented.⁵

The acknowledged medical management options consist of postural nursing, nasopharyngeal tubes,⁶ and endotracheal tubes. Nasogastric tubes have been used in a

handful of cases for the dual purpose of maintaining the airway maintenance and supplementary feeding.⁷ Nasopharyngeal tubes are recommended initially to alleviate the immediate consequences of hypoxia,⁸ but have not been universally successful.⁹ Laryngoscopy for intubation or endoscopic evaluation is often difficult and sometimes impossible.¹⁰ ‘Awake intubation’ without general anaesthesia is safer and less difficult, using a special purpose slotted laryngoscope. Techniques such as passing tracheal guide wires through the suction port of a flexible paediatric bronchoscope have been tried. The bronchoscope is then removed and an endotracheal tube is threaded over the wire. This technique is safe and allows rapid endotracheal intubation in difficult infants. Ultrathin bronchoscopes have been developed, which have provided a means of flexible endoscopic intubation for neonates or small infants with dysmorphic airways and various degrees of micrognathia.

Operations should be considered only when non-invasive medical attempts at management of the airway have failed. These options include glossoptexy, subperiosteal release of the floor of the mouth, and tracheotomy.

Glossoptexy facilitates anterior lingual positioning and is done in infancy. It is reversed at the time of palatal repair, near the end of the first year of life. This period is critical to pre-speech vocalisation. Several studies have shown the success of glossoptexy operations where airway maintenance is concerned. A less commonly used treatment option is subperiosteal release of the floor of the mouth. In rare cases, obstructed airways that are unlikely to improve over a long period may require a tracheotomy. Recent studies show this to be the favoured method of long-term management, although responses from those surveyed were often based on emotions as much as scientific data.¹¹ The mechanism of obstruction obviously has an effect on management, so it has been postulated that the management of the obstruction should be based on the results of direct endoscopic observation of the airway and not on the presumptive diagnosis of glossoptosis. It should not be assumed that ‘catch-up growth’ of the mandible will lead to spontaneous resolution of either the airway obstruction or micrognathia. The presentation of the child with Pierre Robin syndrome should not be the end of the diagnostic search but rather the beginning.¹²

The management of a child with Pierre Robin syndrome requires a multi-disciplinary team approach. Paediatricians, maxillofacial surgeons, orthodontists, specialist nurses, speech therapists, and counsellors are all involved, with each member of the team having a valuable role.

Children with Pierre Robin syndrome present unique management problems and we have shown the effective use of a modified nasopharyngeal tube. The readings from the pulse oximeter provided the medical staff with a continuous indication of oxygenation.

The data can be saved, printed out, and included in the patient's notes. We think that this simple technique may allow earlier discharge of these infants with home monitoring equipment.

REFERENCES

1. Myer CM, Reed JM. Airway management in Pierre Robin syndrome. *Otolaryngol Head Neck Surg* 1998; 118: 630–635.
2. Shinkwin CA, Gibbin KP. Tracheostomy in children. *J R Soc Med* 1996; 89: 188–192.
3. Singer L, Sidoti EJ. Pediatric management of Robin syndrome. *Cleft Palate Craniofac J* 1992; 29 (3): 220–223.
4. Masters IB, Chang AB. Modified nasopharyngeal tube for upper airway obstruction. *Arch Dis Child* 1999; 80: 186–187.
5. Tomaski SM, Zalzal GH, Saal HM. Airway obstruction in the Pierre Robin sequence. *Laryngoscope* 1995; 105 (2): 111–114.
6. Heaf DP, Helms PJ. Naso-pharyngeal airways in Pierre-Robin syndrome. *J Pediatr* 1982; 100: 698–703.
7. Prodochl DC, Shattuck KE. Nasogastric intubation for nutrition and for airway protection in infants with Robin syndrome. *J Perinatol* 1995; 15 (5): 395–397.
8. Singer L, Sidoti EJ. Pediatric management of Robin syndrome. *Cleft Palate Craniofac J* 1992; 29 (3): 220–223.
9. Shinkwin CA, Gibbin KP. Tracheostomy in children. *J R Soc Med* 1996; 89: 188–192.
10. Benjamin B, Walker P. Management of airway obstruction in the Pierre-Robin syndrome. *Int J Pediatr Otorhinolaryngol* 1991; 22 (1): 29–37.
11. Myer III CM, Reed JM, Cotton RT, Willing JP, Shott SR. Airway management in Pierre-Robin syndrome. *Otolaryngol Head Neck Surg* 1998; 118 (5): 630–635.

12. Shprintzen RJ, Singer L. Upper airway obstruction and the Robin syndrome. *Int Anaesthesiol Clin* 1992; 30 (4): 109–114.
13. La glossoptose. Son diagnostic, ses consequences, son traitement. *Bulletin de l'Académie nationale de médecine, Paris* 1923; 89: 37; *J Med Paris* 1923; 43: 235–237.

The Authors

I. S. Whitaker MA, MB, B Chir, Cantab
Demonstrator in Anatomy
Department of Anatomy
University of Cambridge, Cambridge, UK
S. Koron BDS, MFDS
Senior House Officer
Department of Oral and Maxillofacial Surgery
South Manchester University Hospitals NHS Trust,
Manchester, UK
D. W. Oliver MB, BCh, FRCS
Specialist Registrar
Department of Plastic and Reconstructive Surgery
Radcliffe Infirmary, Oxford, UK
P. Jani BDS, MBBS, FRCS (Lond), FRCS (Orl)
Consultant ENT and Head and Neck Surgeon
Addenbrooke's Hospital, Cambridge, UK

Correspondence and requests for offprints to: Dr I. S. Whitaker MA, MB, B Chir, Cantab, Demonstrator in Anatomy, 8 Avonlea Road, Sale, Cheshire M33 4HZ, UK. Tel: +44 (0) 161 9627563; E-mail: iain.whitaker@yahoo.com

Accepted 13 May 2003